## Contents

**Foreword: Pediatric Surgery**  
Ronald F. Martin  

### Management of Anorectal Malformations and Hirschsprung Disease  
Colin D. Gause and Sanjay Krishnaswami  

Anorectal malformations (ARM) and Hirschsprung disease (HD) are managed with placement of normal intestine within the anal sphincter complex. Long-term complications specific to ARM include fistula remnants, recurrence, urinary reflux with associated chronic renal insufficiency, sexual dysfunction, and fertility difficulties. Complications specific to HD include enterocolitis, persistent or acquired aganglionosis, and internal sphincter achalasia. ARM and HD patients are both at risk of stricture, bowel dysfunction and incontinence, which can have a severe impact on quality of life. Bowel management strategies should be tailored to the patient’s specific category of bowel dysfunction.

### Abdominal Tumors: Wilms, Neuroblastoma, Rhabdomyosarcoma, and Hepatoblastoma  
Jennifer T. Castle, Brittany E. Levy, and David A. Rodeberg  

Pediatric cancer patients have improved outcomes over the past several decades leading to a greater number of survivors living well into adulthood. Owing to their increased longevity, adult care providers are encountering childhood cancer survivors with greater frequency in their clinics and hospitals. Childhood cancer treatments are associated with varied and significant systemic complications that either persist or develop well into adulthood, including secondary malignancies, cardiomyopathies, and adhesive disease that can complicate even the simplest operation. This article reviews four of the most common solid abdominal tumors in the pediatric population and the long-term sequelae of their respective treatment regimens.

### Congenital Diaphragmatic Hernia: Considerations for the Adult General Surgeon  
Xiao-Yue Han, Leigh Taryn Selesner, and Marilyn W. Butler  

The contemporary pillars of congenital diaphragmatic hernia (CDH) management include prenatal diagnosis for multidisciplinary care coordination and counseling, medical optimization after birth, and elective (not emergent) operative repair after stabilization, allowing for improvement in pulmonary hypertension and maturation of lungs. Lung hypoplasia and pulmonary hypertension in infants with CDH represent a medical emergency, not one that necessitates immediate surgery. Many infants surviving CDH repair have significant morbidities that may persist into adulthood. Rare cases of previously occult CDH may present acutely in the older child or adult with nonspecific gastrointestinal or pulmonary symptoms.
Esophageal atresia (EA) with tracheoesophageal fistula (TEF) is among the most common congenital anomalies requiring surgical intervention in infancy. General surgeons practicing in rural or austere environments may encounter emergency situations requiring their involvement. Respiratory emergencies can arise in the neonatal period; the recommended approaches are the ligation of the fistula through the chest or occlusion of the distal esophagus through the abdomen. As survivors of the condition reach late adulthood, general surgeons can anticipate encountering these patients. An understanding of risk factors, common symptoms, associated anomalies, and the appropriate diagnostic evaluation will facilitate care.

Pediatric ingestions encompass a wide range of diseases, including foreign body ingestions, caustic ingestions, and aspiration. Specific topics of interest in the pediatric age group for adult general surgeons are button batteries and magnets, which have significant morbidity and mortality and require a high index of suspicion to provide timely care. Evaluation and management of these cases should be tailored to the offending agent and managed at an appropriate pediatric center.

Perforated appendicitis continues to be a significant cause of morbidity for children. In most centers, ultrasound has replaced computed tomography as the initial imaging modality for this condition. Controversies surrounding optimal medical and surgical management of appendicitis are discussed. Management of intussusception begins with clinical assessment and ultrasound, followed by image-guided air or saline reduction enema. When surgery is required, laparoscopy is typically utilized unless bowel resection is required. The differential diagnosis for pediatric gastrointestinal bleeding is broad but often made with age, history, and physical examination. Endoscopy or laparoscopy is sometimes needed to confirm a diagnosis or for treatment.

Congenital abdominal wall defects vary from abdominal wall hernias to severe congenital structural anomalies that include gastroschisis, omphalocele, and prune belly syndrome. The conditions often carry various associated anomalies and require multidisciplinary treatment approaches. Complex surgical reconstructive techniques are frequently required and prenatal, perioperative, and long-term follow-up is critical to ensuring the best possible outcomes.
Small Bowel Congenital Anomalies: A Review and Update 821
Grant Morris and Alfred Kennedy Jr

The small intestine is a complex organ system that is vital to the life of the individual. There are several congenital anomalies that occur and present most commonly in infancy; however, some may not present until adulthood. Most congenital anomalies of the small intestine will present with obstructive symptoms, whereas some may present with vomiting, abdominal pain, and/or gastrointestinal bleeding. Various radiologic procedures can aid in the diagnosis of these lesions that vary depending on the particular anomaly. The congenital anomalies of the small intestine discussed include Meckel diverticulum, duodenal web, duodenal atresia, jejunoileal atresia, and intestinal duplications.

Malrotation: Management of Disorders of Gut Rotation for the General Surgeon 837
Woo S. Do and Craig W. Lillehei

In this article, we aim to provide the general surgeon with a clinical blueprint to navigate disorders of gut rotation. We emphasize that bilious emesis in a newborn is malrotation with volvulus until proven otherwise. Although an upper GI series can establish the diagnosis, surgical intervention should not be delayed until the child is ill-appearing. Following detorsion, the key steps are to broaden the mesentery, fully Kocherize the duodenum, and mobilize the cecum. If nonviable bowel is encountered, the principles of damage control can be applied to children. Every effort should be made to preserve bowel length.

Surgical Support of the Developmentally Delayed or Neurologically Impaired Child 847
Robert L. Ricca and Edward Penn

Children with underlying neurologic conditions or developmental delay may have undergone prior surgical therapy to improve quality of life. These patients may present to the emergency room with complications associated with these procedures or present requiring emergent or urgent surgical management of a new diagnosis. An understanding of the anatomic variation and known long-term complications of these devices is important for any surgeon who may be called to care for these patients. The goal of this article was to provide recommendations that will assist the general surgeon in the surgical management of children with neurologic impairment or developmental delay.

Medical and Surgical Aspects of Intestinal Failure in the Child 861
Danielle Wendel and Patrick J. Javid

Medical and surgical care for children with intestinal failure has evolved so that long-term life expectancy is common even in the setting of the shortest bowel lengths. The long-term administration of parenteral nutrition has become safe with alterations in lipid formulation, and the risk of liver injury has been dramatically reduced. Well-established techniques for bowel lengthening and tapering exist to increase the absorptive capacity of the remnant bowel. These advances allow for ongoing intestinal rehabilitation in the child with the ultimate goal of enteral autonomy while the use of intestinal transplantation in this population has declined in recent years.
Meconium Ileus, Distal Intestinal Obstruction Syndrome, and Other Gastrointestinal Pathology in the Cystic Fibrosis Patient

Joseph Tobias, Mckinna Tillotson, Lauren Maloney, and Elizabeth Fialkowski

Cystic fibrosis is an autosomal-recessive defect in the cystic fibrosis transmembrane conductance regulator (CFTR) gene located on chromosome 7 that affects 1 in 2500 live White births. Defects in the gene lead to abnormally thick secretions causing chronic obstruction in the respiratory and gastrointestinal tracts. Common gastrointestinal pathology in children with cystic fibrosis includes meconium ileus in infancy and distal intestinal obstruction syndrome in childhood and exocrine pancreatic insufficiency, constipation, and rectal prolapse. This article describes the presentation, diagnosis, and management of these conditions in patients with cystic fibrosis, from birth to adulthood.

Chest Wall Deformities and Congenital Lung Lesions: What the General/Thoracic Surgeon Should Know

J. Duncan Phillips and John David Hoover

Pectus excavatum, carinatum, and arcuatum are 3 developmental chest wall deformities that may evolve during childhood and cause cardiac and/or pulmonary compression. Evaluation may include nonsurgical subspecialty consultations and imaging studies. Treatment may be nonoperative or surgical. Long-term follow-up studies have identified rare complications of traditional open repair. Routine in utero ultrasonography has led to increasing identification of congenital lung anomalies, including congenital cystic adenomatoid malformations, pulmonary sequestrations, and bronchogenic cysts. Short-term follow-up studies have suggested that some lesions may regress spontaneously. Minimally invasive techniques, including thoracoscopy, may allow for early surgical resection with less morbidity than traditional open surgery.

Pediatric Inflammatory Bowel Disease for General Surgeons

Michael R. Phillips, Erica Brenner, Laura N. Purcell, and Ajay S. Gulati

Key differences exist in pediatric and adult inflammatory bowel disease (IBD), and a multidisciplinary approach focused on meeting these needs should be implemented. In an emergency situation, surgical management of pediatric IBD should focus on patient stabilization with an eye toward future intestinal function.